Case reports

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Truncus solitarius pulmonalis

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A case of truncus solitarius pulmonalis is reported in a baby girl who lived for 24 hours and in whom an electrocardiogram was recorded. The complete electrocardiogram has not previously been published in this anomaly. A feature of embryological interest, a punctum, present in the left sinus of the truncal valve, is described.

Truncus solitarius pulmonalis is a rare anomaly of the heart. Fifteen cases have been described since 1814, some of which cannot be verified at this distance. The deformity, which is a variant of the hypoplastic left heart syndrome, is identified by the absence of coronary ostia in the sinuses of the valve of the truncus and by an atretic aorta which appears to be an anomalous coronary artery.

Case history

The patient was born at Edgware General Hospital, the second living child of a 29-year-old Indian mother. The first infant was stillborn and the second is alive and well. The patient was the product of the third, normal pregnancy.

The infant weighed 3544 g at birth, and cried spontaneously. At 1 hour she was slightly cyanosed, with shallow, grunting respiration. At 18 hours her condition deteriorated rapidly after feeding, when she became deeply cyanosed with shallow respiration, 30 to 32 per minute, with a feeble cry.

On examination, the patient was hypotonic. The pulse rate was regular at 124/minute. The first and second heart sounds were present, and the femoral pulses were equal in volume. The liver was not palpable.

Laboratory investigations were normal apart from the haemoglobin which was 13·6 g/100 ml.

The patient was given 25 per cent oxygen by inhalation.

At 19 hours, examination showed an arched back and clenched fists. Lumbar puncture was unsuccessful.

A systolic murmur was heard for the first time at 21 hours. The respiratory rate was 60/minute, the liver was felt at two fingers' breadth below the costal margin, and the femoral pulses were barely palpable.

Chest *x*-ray was reported to be within normal limits for a newborn.

The electrocardiogram (Fig. 1) showed sinus tachycardia, rate 165/minute, PR interval 0·14 sec, QRS axis −120°. The right ventricle was abnormally dominant suggesting hypoplasia of the left ventricle.

Digoxin, 0·06 mg/kg and frusemide 5 mg were given intramuscularly.

The infant suffered cardiac arrest at 22 hours, was intubated, and resuscitated with external cardiac massage. Continuous ventilation was required thereafter. She was transferred to another hospital for possible surgical treatment, but died in the ambulance after a second cardiac arrest.

Necropsy

The abdominal viscera were normally related. The adrenal glands were large, but, apart from the heart, the viscera were normal.

The heart was normally sited and of normal size for a newborn. It weighed 19 g. A single arterial trunk arose anteriorly from the right side of the heart, and gave rise to the pulmonary arteries.

The atria were in the normal relation. The venae cavae drained into a large right atrium, and the pulmonary veins were received into a hypoplastic left atrium. The foramen ovale was widely patent.

There was a single ventricle with the morphological characteristics of a right ventricle, which gave rise to the arterial trunk. The trunk was 7·5 mm in diameter. Only one atrioventricular valve with three cusps was present and was not continuous with the truncal valve. Fig. 2 shows the inside of the heart.

The left atrioventricular valve was absent, the only route from the left atrium to the ventricle being through the foramen ovale.

The valve of the truncus was 11 mm in diameter and had three cusps. The left sinus of the truncal valve had a minute punctum (Fig. 2 inset) which did not give rise to a coronary artery.

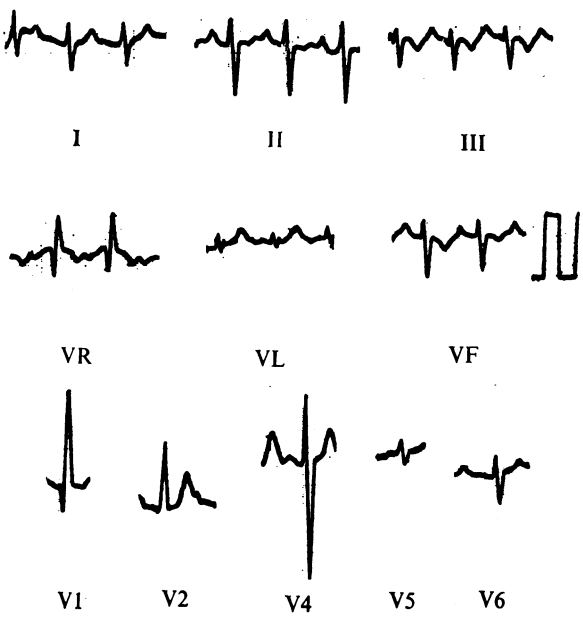

FIG. 1 *The electrocardiogram* (see text).

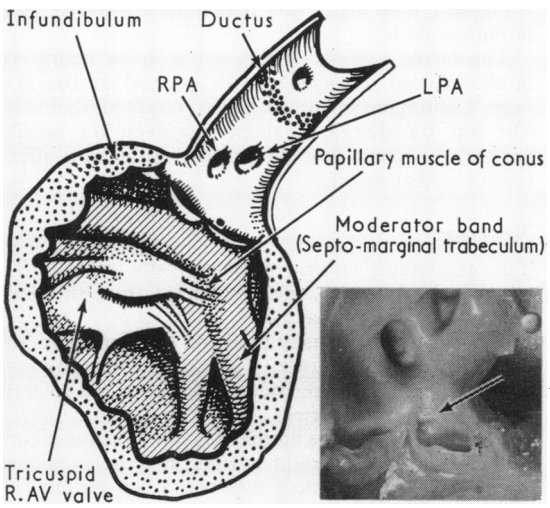

FIG. 2 *Drawing of the ventricle and the truncus. The inset is a photograph of the valve with the punctum in the right aortic sinus* (arrowed).

The right and left pulmonary arteries arose from the right and left posterolateral walls of the truncus, respectively. The right pulmonary artery was 4·5 mm and the left 5 mm in diameter.

The vessels of the arch were abnormally arranged. The innominate artery arose from the arch of the trunk, and from it originated the carotid and subclavian arteries (Fig. 3a). The atretic aorta, 16 mm in length, looked like a coronary artery arising from the innominate artery. This vessel in fact originated from the atrioventricular groove (Fig. 3b). No left ventricular cavity was found, despite careful search. The internal diameter of the atretic aorta was 2 mm. The coronary arteries arose from the three ill-defined sinuses and three valve cusps were discernible with a hand lens.

The pulmonary trunk and the arch of the aorta were joined by a ductus arteriosus, 7 mm in length. The ductus portion was of the same diameter as the truncus, and was recognizable by its wrinkled intimal surface.

Discussion

Truncus solitarius pulmonalis was first described by Farre in 1814. His account of the anomaly is beautifully illustrated with engravings. Forrester's case, in 1847, is very similar to the subject of this communication except that his example had a common atrium and also a family history of congenital heart disease. In her review of truncus arteriosus communis, Humphreys (1932) defined the features of truncus solitarius pulmonalis with considerable emphasis, and indicated that it had been confused with persistent truncus arteriosus and truncus solitarius aorticus. Siddoway and Chernish (1952) tabulated 6 examples from the earlier published reports and added 3 of their own, essentially similar to the case considered here.

Most authors discovered a rudimentary left ventricle from which the atretic aorta arose. The case described by Popjak (1942) from the British Postgraduate Medical School (now the Royal Postgraduate Medical School) had an atretic but recognizable left ventricle. The necropsy report of the case was available to us, but regrettably the specimen was lost in the last war. Despite meticulous search, the present case revealed no subvalvar chamber.

Truncus solitarius pulmonalis differs from other forms of truncus and is characterized by a single *right* ventricle, a solitary atrioventricular valve, and a grossly abnormal coronary circulation; the sinus of the truncal valve is usually devoid of coronary ostia.

This subject had three coronary arteries, one arising from each of three sinuses of the atretic aorta, and a punctum in the left sinus of the truncal valve. This had not previously been described. Von Konstantinowitch's (1906) case (quoted by Humphreys, 1932) had one coronary artery arising from a truncal sinus. The punctum in the present subject probably represents an underdeveloped coronary ostium. Anomalous coronary arteries, arising from the pulmonary artery are rare: Roberts (1962), reviewing that finding, quotes Hackensellner's (1956) observation of 6 coronary anlagen in the sinuses of the embryonic great arteries. Thus, a

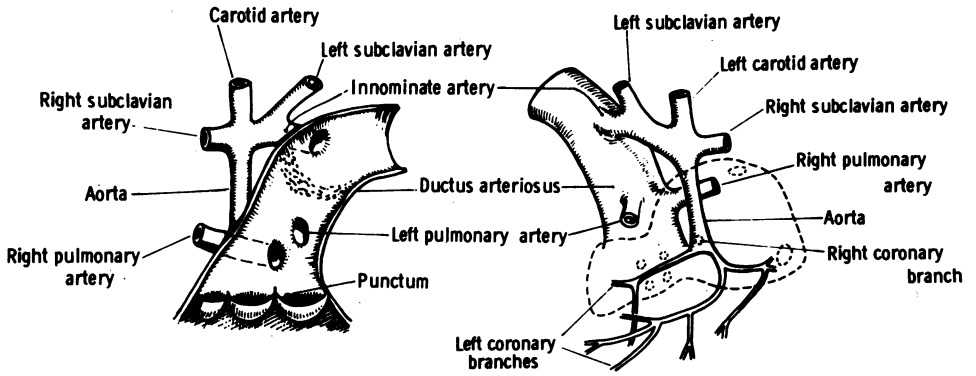

FIG. 3 Left: *Anterior aspect of the truncal valve, the ductus arteriosus, and the interrupted aortic arch.* Right: *Posterior view of the truncus to show the distribution of the coronary arteries. The atria and their veins are indicated in dotted line.*

potentiality exists for a coronary artery to arise from any of the six sinuses of the great arteries.

It seems likely that truncus solitarius pulmonalis is due to defective development of the bulbar ridges very early in embryonic life. If the bulbar ridges form too far posteriorly in the bulbus cordis, the truncus arteriosus will be partitioned by the spiral septum at the expense of the aorta. This theory is supported by the findings of de la Cruz and da Rocha (1956).

In truncus solitarius pulmonalis the rudimentary aorta usually lies behind the common pulmonary trunk, though Blake *et al.* (1964) illustrated an example in which the aorta lies in front.

Unlike transposition of the great arteries, which may be associated with a single ventricle, truncus solitarius pulmonalis is always associated with a single ventricle. Wilson (1798) depicted a posteriorly placed truncus in the case of ectopia cordis with a biloculate heart. The coronary distribution was not described, but his account suggests that the truncus was a pulmonary trunk.

Van Praagh and Van Praagh (1965) described truncus solitarius pulmonalis as pseudotruncus pulmonicus, and restated that it was not related to common aorticopulmonary trunk (truncus arteriosus communis). Whether truncus solitarius pulmonalis should be defined as a pseudotruncus is open to discussion. Lev and Saphir (1942) defined a truncus as the only great artery arising from the base of the heart, giving rise to the coronary, pulmonary, and systemic circulations. A solitary pulmonary trunk fulfils these criteria, though the coronary arteries are perfused in the retrograde direction through the ductus arteriosus.

So far as can be ascertained, none of the published cases of truncus solitarius pulmonalis had a functional left ventricle, and none had two atrioventricular valves.

'Aortic and mitral atresia' has been used to describe truncus solitarius pulmonalis. While true, the description is imprecise. The subject of this communication had no evidence of development of any part of the left atrioventricular valve, which suggests a developmental failure of the left lateral and left superior endocardial cushions; the term mitral agenesis is thus more precise.

Truncus solitarius pulmonalis is a recognizable cognomen for the anomaly, and has been used for many years. It seems reasonable, therefore, to continue to apply it to those subjects who exhibit the anatomical features described.

All the previous reports indicate that like all hypoplastic left heart syndromes, truncus solitarius pulmonalis has a very poor prognosis.

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